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journal homepage: www.jpascasereports.comPediatric Crohn disease complicated by an entero-uracho-cutaneous fistula[☆]

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ABSTRACT

Crohn disease is characterized by transmural inflammation and can thus lead to fistulization between the inflamed bowel and adjacent structures, typically other loops of bowel, skin, or bladder. Rarely, fistulae to embryonic remnants such as a patent urachus may also occur. This case report describes an eleven year-old female patient with a new diagnosis of Crohn disease who presented with an entero-uracho-cutaneous fistula. The unique challenges to both diagnosis and management in this patient population are discussed.

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Fistulizing Crohn disease can present significant clinical challenges. One example is the development of a fistula between bowel and the urachus, an embryonic remnant. Only a few cases exist in the literature. Our case describes a pediatric patient with Crohn disease who presented with an entero-uracho-cutaneous fistula, and, what we regard as an optimal approach for the management of this problem.

1. Case report

The patient was an eleven year-old female who initially presented with a one month history of recurrent fever, abdominal pain, and diarrhea. Abdominal tenderness was present particularly in the right lower quadrant. The white blood cell count, erythrocyte sedimentation rate, and C-reactive protein were all elevated. An initial abdominal ultrasound indicated evidence of thickened and presumably inflamed small bowel loops in the right lower quadrant. A presumptive diagnosis of Crohn disease was made. Upper and lower gastrointestinal endoscopy were non-diagnostic, therefore, a magnetic resonance (MR) scan was done.

This confirmed inflammation of the terminal ileum and also showed an incidental finding of an urachal cyst ([Fig. 1](#)). The interpreting radiologist noted thickening and apparent inflammation of the urachal cyst as well but stated that it was unclear as to whether this was a consequence of the adjacent small bowel disease.

One month later the patient was admitted to hospital because of the development of purulent, bloody, brown drainage from the umbilicus. An ultrasound revealed a fistulous connection between the urachal cyst and the small bowel ([Fig. 2](#)). The Pediatric Surgical service was consulted and we elected to bridge her to surgery in collaboration with the Pediatric Gastroenterologists with non-operative/medical management initially which consisted of intravenous broad-spectrum antibiotics, oral prednisone, and restricting her to a low residue semi-elemental diet. Over a period of six weeks, good results were achieved with resolution of her fevers and abdominal pain, as well as significantly reducing her umbilical discharge. A complete steroid taper was achieved within weeks, before surgery.

Laparoscopy confirmed the diagnosis of Crohn disease with the finding of ileal "creeping fat" ([Fig. 3](#)) and was followed by a laparoscopic-assisted ileocecal resection of all grossly affected bowel along with the urachal cyst and bladder dome via a minimal laparotomy. Resection was completed in one stage with a primary bowel anastomosis and over-sewing of the bladder. The pathology of the resected segments was consistent with Crohn disease of the ileum and an inflamed urachal cyst. The patient made a quick recovery and was discharged on post-operative day 4 on

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Fig. 1. Sagittal view MR image shows the cystic urachal remnant (arrow) with fistulous communication to umbilicus juxtaposed to a thickened small bowel segment.

metronidazole and azathioprine. The patient suffered no operative complications and is now thriving and asymptomatic on only a low dose of azathioprine at two years post-bowel resection.

2. Discussion

An urachal cyst, or sinus, can form when the urachus fails to obliterate into the median umbilical ligament during gestation. This embryonic remnant of the allantoic duct resides in the space of Retzius and connects the bladder to the umbilicus. The incidence is estimated at 1-in-5000 [1]. These urachal anomalies, usually as cystic lesions, can become infected, undergo malignant transformation, or cause complications such as fistulae. Therefore, removal is typically advised when symptomatic [2,3].

Crohn disease is characterized by transmural inflammation and can thus lead to fistulization between the inflamed bowel and adjacent structures. Crohn fistulae are most commonly between other loops of bowel or the skin. However, fistulae are also known

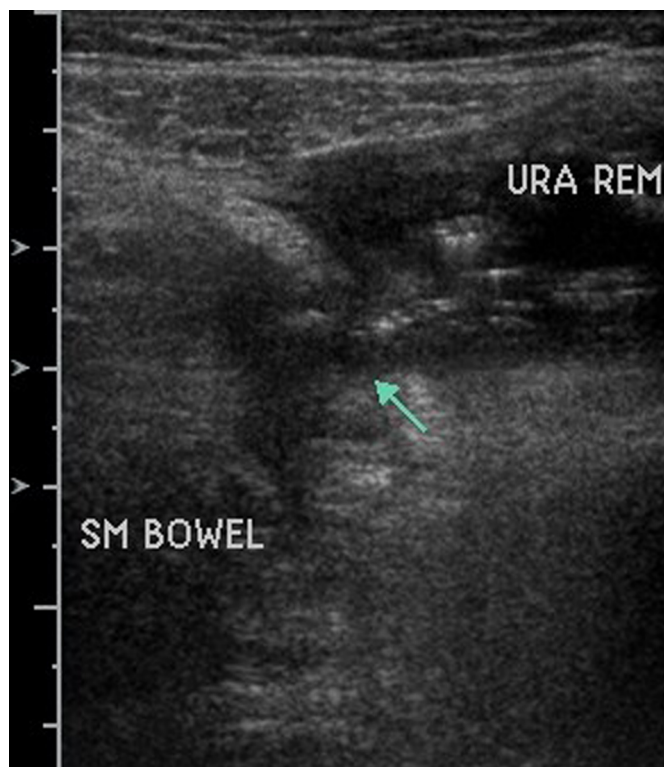


Fig. 2. Ultrasound image demonstrating a fistulous communication (arrow) between a loop of small bowel and the urachal cyst.

to occur with structures of the genitourinary tract, estimated at approximately 18% of operative cases [4].

Our case describes a unique complication of Crohn disease in which a fistula developed between inflamed terminal ileum, an urachal cyst, and the umbilicus. Only a handful of similar cases exist in the English literature [5–8], while ours is the youngest patient described to date. A female patient just eleven years of age presents unique challenges in both diagnosis and management. The most

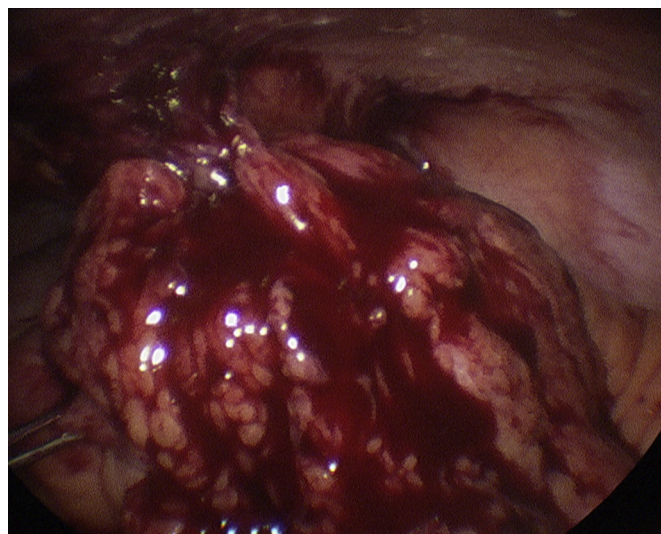


Fig. 3. Laparoscopic photograph demonstrating an entero-urachal fistula on the left side of the image. Creeping fat on an adjacent loop of small bowel can be seen to the right.

comparable previously reported case published in 1980 involved a 28 year old male patient [5].

Diagnosis begins with a thorough history and physical examination, looking for signs and symptoms of Crohn disease and the associated complications. In the case of feculent drainage from the umbilicus, the possibility of an external Crohn fistula should be immediately suspected and investigated with appropriate imaging studies. However, fistulae secondary to diverticulitis can present in a similar manner [9], although this is not a diagnosis that would normally be entertained in a child. Also, purulent or serous umbilical drainage likely represents a primary urachal infection.

Previous reports have utilized and recommended computed tomography scanning to establish a diagnosis in these complicated fistulae cases [7,8]. However, this did not seem prudent in our case due to radiation-related risks [10]. Fortunately, we were able to successfully diagnose the nature of the fistula using a combination of ultrasound and MR. Other reports have also described the use of contrast enhanced x-rays or barium studies [6,7]. Though these were not necessary in our case, a sinogram performed through the umbilicus would have been a logical next step if the diagnosis was still in doubt.

This case also highlights the usefulness of pre-operative medical management in the treatment of complicated Crohn fistulae. The use of antibiotics and steroids helped to settle the inflammation and allows a safer, single-staged operation [11]. A complete steroid taper was achieved pre-operatively. Oral nutritional support has been advocated, even as primary treatment, and was successfully employed in our case as supplemental therapy with good results [12,13].

The use of infliximab, an anti-tumor necrosis factor monoclonal antibody, has also been described in the medical management of fistulizing Crohn disease and has shown benefit as both induction and maintenance therapy in adult trials [14], with similar results observed in the pediatric population [15]. This could have been considered in our case if initial medical management had failed to respond to steroids. However, resection of the infected urachus would still be required and significant cost is associated with this medication.

Previous reports have advocated for complete *en bloc* resection of the fistulous segment, including the involved bladder dome. In our case we were able to accomplish this using a laparoscopic-assisted approach, limiting the size of the abdominal incision. This is contrary to previous reports which describe formal laparotomy. Bowel mobilization was completed laparoscopically, followed by a small lower midline incision to resect and anastomose the bowel, then resect the urachus and bladder dome. Pre-operative medical

therapy limited inflammation in the area to allow such a resection, limit incision length, and decrease complication risk. In our case an excellent outcome was achieved using current techniques for both diagnosis and management.

Consent

Written informed consent was obtained from the patient's guardian for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest statement

We have no conflicts to disclose.

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